Chapter 27

Cardiology

History

- In newborns, exercise tolerance is approximated by asking caregivers about feeding difficulties, specifically tachypnea, cyanosis, and diaphoresis during feeding
- In infants, failure to thrive (gain weight at an appropriate rate) warrants consideration of congenital heart disease
- Children with other congenital anomalies are more likely to have congenital heart disease
  - Particularly defects along the midline or involving other solid organs
  - Family history should focus on family members with heart disease (congenital or acquired), sudden unexplained deaths, and arrhythmias

Examination

- Blood pressure should be measured in both arms and one leg to evaluate for coarctation of the aorta
- Murmurs: more than 90% of all children are noted to have a murmur at some time in their lives
  - Most murmurs are harmless
  - Characteristics concerning for a pathologic murmur include associated cardiac symptoms, a loud or harsh-sounding systolic murmur (> 3/6 or with a palpable thrill), a diastolic murmur, abnormal heart sounds, presence of a click, and weak or absent peripheral pulses
- Electrocardiograms (ECGs) are helpful if available (consult *The Harriet Lane Handbook* for help interpreting an ECG)

Evaluating the Cyanotic Newborn, Infant, and Child

- Central cyanosis is universally consistent with hypoxemia and is best appreciated in the oral mucosa, conjunctivae, and the tip of the tongue

- The most common cause of cyanosis in newborns, infants, and children is respiratory compromise from a host of pulmonary diseases
- Cyanosis from congenital heart disease is typically due to a right-to-left intracardiac shunt and persists beyond the newborn period
- Evaluation
  - Initial evaluation should include pulse oximetry and measurement of the partial pressure of oxygen in arterial blood (PaO_2) by blood gas to confirm hypoxemia
    - The PaO_2 in a normal, 1-day-old newborn may be as low as 60 mmHg
  - Primary pulmonary processes are associated with tachypnea and dyspnea; congenital cyanotic heart defects are generally associated with effortless tachypnea often described as "comfortable tachypnea"
    - If a cyanotic heart defect is suspected, perform chest radiograph, ECG (if available), and a hyperoxitest
    - Ultimately, the diagnosis of congenital cyanotic heart disease is made by echocardiography
    - The hyperoxitest compares PaO_2 and pulse oximetry values after a 100% oxygen challenge as a means of differentiating pulmonary from cardiac causes of hypoxemia
      - Arterial blood gas samples should be taken from the right upper extremity
      - PaO_2 and pulse oximetry values are taken while the newborn is breathing fraction of inspired oxygen (FiO_2) = 0.21, and after FiO_2 is increased to 1.00 for 10 minutes
      - Care should be taken to get the FiO_2 as close as possible to 1.00
        - Pulmonary disease: an increase in $PaO_2 > 150$ and pulse oximetry = 100%, with $FiO_2 = 1.00$
        - Cardiac disease: $PaO_2 < 150$ and pulse oximetry < 90%, with $FiO_2 = 1.00$
- Treatment
  - Although some children with cyanotic heart defects can survive past infancy (eg, most commonly those with mild

forms of Tetralogy of Fallot), definitive treatment of cyanotic heart defects requires surgical correction
    - In the newborn period, if surgical intervention is a feasible option, a prostaglandin E_1 intravenous (IV) infusion can be started to maintain patency of the ductus arteriosus
        - The starting dose of prostaglandin E_1 is 0.05–0.1 μg/kg/min
        - Common side effects include flushing, apnea, hypotension, and fever

Arrhythmias

- Bradycardia
    - Most common cause of true bradycardia in infants and children is hypoxia, usually due to respiratory compromise
    - Sinus bradycardia can be normal in adolescents and athletes
    - Can also be caused by increased vagal tone, increased intracranial pressure, hyperkalemia, hypercalcemia, hypothyroidism, hypothermia, long QT syndrome, and drugs (eg, digoxin, β-blockers)
    - In newborns, physiological stresses (eg, hypoxia, cold, hypoglycemia) often manifest as bradycardia (manifest as tachycardia in older children and adults)
    - Bradycardia can also occur as a result of heart block
    - Management
        - Treat the underlying cause
        - A hemodynamically unstable patient with underlying bradycardia requires emergent attention
        - Begin with effective oxygenation and ventilation via bag-valve mask if necessary
        - Epinephrine is the drug of choice after oxygen; dose 0.01 mg/kg (0.1 mL/kg) of 1:10,000
        - Atropine should be considered a second-line agent, unless vagal stimulation is thought to be the source of the bradycardia
            - Dose 0.02 mg/kg (minimum dose 0.1 mg)
- Tachycardia
    - Most common cause of tachycardia in pediatric patients is sinus tachycardia

        - This can be caused by hypovolemia, hemorrhage, hypoxia, anemia, fever, sepsis, shock, congestive heart failure, myocardial disease, anxiety, and drugs (eg, β-agonists, atropine)
    - Management requires distinguishing sinus tachycardia from supraventricular tachycardia (typically narrow QRS complex) and ventricular tachycardias (wide QRS complex)
        - Sinus tachycardia is almost always accompanied by a history that explains it (ie, volume loss, fever, hemorrhage, etc)
        - Maximum heart rates for infants and children can be surprising
            - Infants: up to 200 beats per minute (bpm)
            - Children: 180 bpm
            - Adolescents: 160 bpm
        - Treatment involves correcting the underlying causes
    - Supraventricular tachycardia is the most common tachyarrhythmia seen in children with increased ventricular rates (infants > 230 bpm, children > 180 bpm)
        - P waves, if visible, are usually abnormal
        - Heart rate is regular, rapid, and monotonous, with minimal variation
        - Narrow QRS complexes are typical
        - Can be associated with congenital heart disease (eg, Ebstein's anomaly, transposition) and preexcitation syndromes, like Wolff-Parkinson-White syndrome
        - Most often idiopathic
        - Treatment includes vagal maneuvers and adenosine (initially 0.1 mg/kg/dose) IV administered quickly, followed immediately by a flush of 5–10 cc normal saline, given its short half-life
        - If patient is unstable, perform synchronized cardioversion (0.5–1 J/kg)
- Atrial flutter
    - Less common and usually associated with a narrow complex tachycardia (unlike in adults) secondary to excellent conduction through the atrioventricular node
    - Treatment includes digoxin, synchronized cardioversion, or overdrive pacing; short-acting β-blockers may be used

to control the ventricular rate
  - Treat the underlying etiology when possible
- Atrial fibrillation
  - Less common in pediatric patients than in adults
  - Defined as an irregular-appearing and fast atrial rate (350–600 bpm), narrow QRS complexes, and an irregular ventricular response rate of 110–150 bpm
  - Causes and treatment are similar to atrial flutter
  - Anticoagulation may be necessary if present for more than 48 hours and pharmacologic or electrical cardioversion can be postponed
- Ventricular dysrhythmias
  - Premature ventricular contraction (PVC)
    - Typically benign; multifocal PVCs are more concerning
    - Causes include myocarditis, cardiomyopathy, congenital and acquired heart disease, long QT syndrome, hypokalemia, hypoxemia, hypomagnesemia, anxiety, and drugs (eg, digitalis, catecholamines, caffeine, and anesthetics)
    - Treatment is only necessary if the PVCs are associated with symptoms, hemodynamic changes, or underlying heart disease, or are made worse with exercise
    - Treatment can include lidocaine, β-blockers, and other antiarrhythmic drugs
- Ventricular tachycardia (VT) and ventricular fibrillation (VF)
  - Uncommon in children
  - Represent the initial arrest rhythm in only 10% of pediatric cardiopulmonary arrests, but will appear during up to 25% of all pediatric codes
  - For stable VT, consider a slow load of amiodarone 5 mg/kg, but be prepared to cardiovert or defibrillate if necessary
  - For pulseless VT and VF, treat with immediate cardiopulmonary resuscitation, defibrillate 2–4 J/kg, and administer either lidocaine (1 mg/kg) IV or amiodarone (5 mg/kg) IV bolus

Congestive Heart Failure

- Congestive heart failure (CHF) can be caused by a variety of congenital or acquired medical conditions

- By far, the most common cause of CHF in infancy is congenital heart disease
  - Volume overload lesions, such as ventricular septal defect, patent ductus arteriosus, and endocardial cushion defects are most common
  - Tachyarrhythmias and heart block can cause heart failure at any age, including in utero
  - Acquired heart diseases become more common as children get older
    - These conditions include myocarditis, acute rheumatic carditis, rheumatic valvular diseases with significant mitral or aortic regurgitation, dilated cardiomyopathy, metabolic abnormalities, endocrinopathies, and severe anemia
- Diagnosis
  - Historically, infants present with poor feeding, tachypnea that worsens during feeding, poor weight gain, and diaphoresis, particularly with feeding
  - Older children report dyspnea (particularly with activity), orthopnea, easy fatigability, and swelling of the eyelids, feet, and hands
  - Like in adults, the following are common physical examination findings consistent with but not specific to CHF:
    - Wheezing or crackles
    - Tachycardia
    - Gallop rhythm
    - Displaced point of maximal impulse
    - Weak pulses
    - Hepatomegaly
    - Extremity and eyelid edema
  - Unlike in adults, jugular venous distention is not a common finding
  - Cardiomegaly is almost always seen on a chest radiograph
- Management
  - Address the underlying etiology, provide support, and control underlying heart failure state
  - Supportive measures include providing adequate calories given fluid restrictions (for infants, as much as 150–169 kcal/kg/day)

- Medical management includes a combination of inotropic agents, diuretics, and afterload-reducing agents

Acquired Heart Disease

- Rheumatic heart disease
  - Acute rheumatic fever is a common cause of heart disease in underdeveloped countries
  - Pathophysiology includes a postinflammatory reaction affecting the whole body
    - Believed to be immunologically mediated following a group A streptococcal infection of the pharynx
    - Other streptococcal infections, like impetigo, do not cause acute rheumatic fever
  - Most cases occur in children between 6–15 years old (peak incidence at 8 y) following a history (1–5 w) of streptococcal pharyngitis with nonspecific symptoms of malaise, fatigue, abdominal pain, and pallor; there is often a positive family history of rheumatic fever
  - The diagnosis is ultimately made using the revised Jones Criteria and requires two major manifestations or one major and two minor manifestations, in addition to evidence of an antecedent streptococcal pharyngitis by either throat culture or serology
  - Treatment includes administering benzathine penicillin G (0.6–1.2 million units intramuscular [IM]) followed by penicillin prophylaxis, relative bed rest, and antiinflammatory therapy
    - Supportive care measures include diuretics, digoxin, morphine, sodium and fluid restriction, and prednisone for severe carditis
    - Preventing recurrence includes administering benzathine penicillin G (1.2 million units IM every 28 days)
      - Alternatively, use penicillin VK 250 mg by mouth (PO) bid or erythromycin 250 mg PO bid for penicillin-allergic patients
      - Prophylaxis should continue until age 21–25 years
- Kawasaki disease
  - Kawasaki disease is an acute, febrile vasculitis of unknown etiology with a predilection for the coronary arteries

- Most common cause of acquired heart disease in developed countries and seen almost exclusively in children < 8 years old
- Diagnosis is clinical and based on a history of high fever lasting 5 days or more, plus four of the following five criteria:
  - Bilateral bulbar conjunctivitis without exudates
  - Erythema of the mouth and pharynx, strawberry tongue, or red and cracked lips
  - Polymorphous exanthema
  - Swelling of the hands and feet, with erythema of the palms and soles
  - Cervical lymphadenopathy (> 1.5 cm), usually single or unilateral
- Additional associated features include extreme irritability, abdominal pain, vomiting, diarrhea, and skin changes (particularly desquamation of the hands, feet, and perineal region), usually after the fever ends
- Laboratory findings include elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), leukocytosis with left shift, normocytic or normochromic anemia, thrombocytosis, sterile pyuria, and elevated liver enzymes
- If left untreated, there is a 15%–25% risk of coronary artery aneurysm and subsequent coronary artery thrombosis and stenosis, although aneurysms can develop elsewhere in the body
- Rarer complications include carditis, valve regurgitation, pericardial effusion, and CHF
- Treatment
  - One dose of IV immunoglobulin (IVIG; 2 g/kg over 12 h)
  - High-dose aspirin (80–100 mg/kg/day divided qid) PO until the fever resolves, followed by 3–5 mg/kg/day of aspirin PO divided once a day for 6–8 weeks, or until the ESR and platelet count return to normal

Infective Endocarditis Prophylaxis

- The use of antibiotic prophylaxis to prevent bacterial endocarditis was greatly changed in 2007. Under current guidelines, prophylaxis is recommended for only the following procedures:

- Dental procedures that involve manipulation of gingival tissues, the periapical region of teeth, or perforation of oral mucosa
- Procedures on the respiratory tract involving incision of the respiratory tract mucosa
- Procedures on infected skin, skin structures, or musculoskeletal tissue
- Cardiac conditions associated with the highest risk of adverse outcome from endocarditis, including:
  - Prosthetic cardiac valve
  - Unrepaired congenital heart disease, including palliative shunts and conduits
  - Congenital heart defects completely repaired with a prosthetic material or device (placed surgically or by catheter) during the first 6 months after the procedure
  - Repaired congenital heart defects with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device
  - Cardiac transplantation recipients with cardiac valvulopathy
  - Previous infective endocarditis
  - Prophylaxis is no longer recommended for procedures involving the gastrointestinal or genitourinary tracts

- Medication and dosing
  - Amoxicillin: 50 mg/kg PO initially (do not exceed maximum adult dose of 3 g), then 25 mg/kg in 6 hours (do not exceed 1.5 g)
  - Ampicillin: 50 mg/kg IV initially, then 25 mg/kg
  - Clindamycin: 10 mg/kg IV initially, then 5 mg/kg
  - Erythromycin ethylsuccinate and stearate: 20 mg/kg PO initially, then 10 mg/kg
  - Gentamicin: 1.5 mg/kg IV initially, then 1 mg/kg
  - Vancomycin: 20 mg/kg IV initially, then 10 mg/kg

Cardiac Syncope

- Although syncope is more frequently the result of vasovagal and orthostatic mechanisms, cardiac causes of syncope include:
  - Obstructive heart lesions
    - Aortic stenosis

        - Pulmonary stenosis
        - Hypertrophic obstructive cardiomyopathy
    - Coronary artery abnormalities
    - Arrhythmias
- Red flags during the evaluation of syncope include:
    - Exercise-induced syncope
    - Preceding chest pain
    - Associated seizure-like activity
    - Atypical history
    - Recurrent or progressive syncope
    - Physical examination suggestive of cardiac disease
    - Abnormal ECG
    - Family history of unexplained death

Chest Pain and Myocardial Infarction

- Chest pain is a frequent chief complaint among children and adolescents, but it is rarely due to any underlying cardiovascular cause. The most common etiologies of chest pain among pediatric patients are:
    - Chest wall pathology
        - Muscle strain
        - Trauma
        - Costrochondritis
    - Respiratory conditions
        - Pneumonia
        - Cough
        - Asthma
    - Gastrointestinal disease, specifically esophagitis related to gastroesophageal reflux disease
    - Psychogenic (anxiety)
- Red flags for underlying cardiovascular disease include:
    - Pain that radiates down the left arm or up the neck, jaw, or back
    - Associated presyncope or syncope
    - Palpitations
    - Dyspnea
    - Dull, squeezing, or heavy chest pain (as opposed to sharp or pinpoint pain that is often reproducible and worse on palpation)

- Cardiac causes of chest pain include:
  - Obstructive congenital heart conditions that put additional strain on the myocardium, such as aortic stenosis, subaortic stenosis, coarctation of the aorta, and pulmonary stenosis
  - Mitral valve prolapse
  - Cardiomyopathy
  - Coronary artery abnormalities
  - Aortic dissection or aneurysm
  - Pericarditis
  - Myocarditis
  - Arrhythmias

Myocardial Infarction

- Rare in children
- Predisposing conditions include history of Kawasaki disease, anomalous origin of a left coronary artery, congenital heart disease, and dilated cardiomyopathy
- Diagnosis and management are similar to that used in adults

Further Reading

1. Robertson J, Shilkofski N. *The Harriet Lane Handbook: A Manual for Pediatric House Officers*. 17th ed. Philadelphia, Pa: Elsevier Mosby; 2005.
2. Park MK. *Pediatric Cardiology for Practitioners*. 4th ed. St Louis, Mo: Mosby; 2002.
3. Keane JF, Lock JE, Fyler DC. *Nadas' Pediatric Cardiology*. 2nd ed. Philadelphia, Pa: Saunders Elsevier; 2006.